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References

[1] Jewett TC, Seigal M. Hiatus hernia and gastro-oesophageal reflux. *J Pediatr* 1984;**3**:340–5.

[2] Cohen S. Motor disorders of the oesophagus. *N Engl J Med* 1979;**307**:184–92.

[3] Eliasson S. Activation of gastric motility from the brain stem of the cat. *Acta Physiol Scand* 1953;**30**:199–214.

[4] Paganini FD, Norman WP, Kasbekar DK, Gillis RA. Effects of stimulation of nucleus ambiguus complex on gastro-duodenal function. *Am J Physiol* 1984;**246**:253–62.

[5] Wood JR, Camilleri M. Low PA, Malagelada JR. Brain stem tumor presenting as an upper gut motility disorder. *Gastroenterology* 1985;**89**:1411–4.

[6] Johnson HD. Active and passive opening of the cardia and its relation to the pathogenesis of hiatus hernia. *Gut* 1966;**7**:392–401.

[7] Hoffman HJ, Becker L, Craven MA. A clinical and pathologically distinct group of benign brain stem gliomas. *Neurosurgery* 1980;**7**:243–8.

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Prognosis in Guillain-Barré, syndrome

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SUMMARY Clinical recovery started in 23 children with Guillain-Barré syndrome after a mean time of 19 days and to full recovery in 15 was 6·2 months. Long term follow up indicated that 19 with acute onset had completely recovered clinically and three with subacute onset had a chronic relapsing or protracted course.

Guillain-Barré syndrome was described more than a century ago, but little has been written about the long term outcome in children. Subacute presentation, severe or prolonged illness, profound sensory loss, papilloedema, CSF pleocytosis, gross slowing of nerve conduction, and onion bulb formation on nerve biopsy are associated with incomplete recovery.[1–3] Most studies deal with adult patients. Eberle *et al*, however, referring specifically to Guillain-Barré syndrome in childhood, found that if 16 days elapsed without improvement after the weakest point was reached there was a 96% probability that recovery would not occur, and if the plateau before improvement lasted longer than 18 days incomplete recovery was almost certain.[2] Because the patients in that study were followed up for a fairly short time and because our experience has been quite different we report the long term outcome in a well defined group of children.

Patients and methods

We studied patients admitted to our hospital during the period January 1970 to December 1985. They were selected according to recommended clinical criteria: progressive motor weakness affecting more than one limb with or without cranial nerve involvement, arreflexia, and absence of pleocytosis.[4] Twenty four patients (13 male and 11 female aged from 19 months to 13 years (mean age 4 years 7 months)) fulfilled the criteria.

Results

Twelve patients had had a preceding illness or event, upper respiratory tract infection being the most common (seven cases); the others comprised measles (two cases), mumps (one), infectious mononucleosis (one), and pneumoencephalography for suspected hydrocephalus (one).

Twenty one patients presented acutely (symptoms for three weeks or less before maximum deficit) and three subacutely (symptoms for six weeks or more before maximum deficit). Three patients who presented acutely had the Miller-Fisher variant.[5] At the time of maximum neurological deficit 10 patients suffered muscle weakness in both arms and legs. This was severe in four, who also had weak diaphragms. Only one patient required ventilation. Eight patients had evidence that the cranial nerves were affected (facial palsy alone in three and multiple cranial nerves affected in five). Six had evidence of autonomic neuropathy manifested as hypertension (three cases), hypotension (one), cardiac arrhythmia (three), profuse sweating (two), and sphincter disturbance (two). The cerebrospinal fluid (CSF) was examined in all patients at least once. A raised CSF protein concentration (mean 1·6 g/l, range 0·1–6·0 g/l) in the absence of a cellular response (less than 10 cells/field) was found in 22

patients. Two patients had a normal CSF protein concentration, but each had only one examination. Treatment consisted of intensive physiotherapy. One child received corticosteroids, and no patient was treated by plasmapheresis.

One patient (aged 19 months) died from cardiorespiratory arrest due to severe autonomic dysfunction. The time taken from maximum deficit to onset of clinical recovery in the remainder ranged from five to 46 days (mean 19·3 days). Time to full clinical recovery (defined as return to normal activities with normal muscle power and return of deep tendon reflexes) was from 2·5 to 15 months (mean 6·2 months) in 15 cases. Of the remaining patients, six were lost to immediate follow up and two were still arreflexic when discharged from hospital.

Twenty two children were followed up and reexamined. The mean follow up interval was seven years for those presenting during 1970–85 and 9·7 years for those presenting during 1970–80. Nineteen were normal on neurological examination and had not developed any complications. Of the remainder, two had chronic relapsing disease and one a protracted illness. The 19 children who had made a full clinical recovery had all originally presented acutely, whereas the three who presented subacutely had a complicated outcome. One patient presented at 13 years of age having had symptoms for three months. She made an incomplete recovery and had two subsequent relapses when aged 16 and 21. At 28 she had persistent distal weakness in the legs, reduced vibration and proprioception, and absent ankle jerks. Another patient presented at the age of 4 having had symptoms for eight weeks. He recovered in three months but relapsed with distal leg weakness, paraesthesia, and arreflexia three years later. He recovered again, and was normal on neurological examination at the age of 10. The third patient had had symptoms for over three months. She improved slowly but was still undergoing rehabilitation one year after presentation. She was unable to walk independently and had mild weakness of the arms, arreflexia, and sensory deficits in the hands and feet.

Discussion

With the eradication of poliomyelitis Guillain-Barré syndrome has emerged as the most common cause of acute motor paralysis in children. It has been proposed that the syndrome in childhood has a different natural history and a better prognosis than that in adults.[1] The few available paediatric studies have reported small numbers of patients, who were often selected or reviewed over short periods of time. The children reported in the study of Eberle *et al* consisted of patients referred to a rehabilitation institution.[2] Only one of our patients, who had a protracted illness, required referral to a rehabilitation centre. The total number of patients in our study, although small, is comparable with that in other studies in children. The age and sex distribution, preceding events, and type and severity of disease were similar to those described by others.[2 6] Treatment was supportive in all patients, with emphasis on padding exposed surfaces to prevent secondary pressure neuropathy. This complication may account for some of the residual deficits in patients whose symptoms and signs are prolonged.[3] One child in our study was treated with corticotrophin, although no benefit was apparent.

On long term follow up all patients who presented acutely had made a complete clinical recovery, although two of these had residual deficits for up to two years. This may account for differences in sequelae in studies conducted over shorter periods. All of the patients who presented subacutely developed chronic complications. Other studies have supported this finding and have shown that a longer time (usually more than three months) to maximum deficit is associated with a risk of relapse or of a protracted course.[3 4] Patients with chronic relapsing or progressive neuropathy seem to have a variant of Guillain-Barré syndrome that can be defined only by the temporal evolution of the disorder.[4] The mean time to onset of recovery from maximum deficit was 19·3 days in our patients. This was longer than that reported by Eberle *et al*.[2] In addition we found no correlation between the rate of recovery or the severity of the illness and the long term outcome.

We conclude that clinical recovery from acute Guillain-Barré syndrome can be expected in all children provided respiratory failure and autonomic neuropathy are appropriately managed. Recovery may not begin for up to six weeks, and full clinical recovery may take up to seven months after the maximum deficit has occurred. Furthermore, subacute presentation seems to indicate a risk of either chronic relapse or a protracted course.

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References

[1] Winer JB, Hughes RAC, Greenwood RJ, *et al*. Prognosis in Guillain-Barré syndrome. *Lancet* 1985;i:1202–3.
[2] Eberle E, Brink J, Azen S, White D. Early predictors of

incomplete recovery in children with Guillain-Barré polyneuritis. *J Pediatr* 1975;**86**:356–59.
[3] Eisen A, Humphreys P. The Guillain-Barré syndrome. *Arch Neurol* 1974;**30**:438–43.
[4] Asbury AK. Diagnostic considerations in Guillain-Barré syndrome. *Ann Neurol* 1981;**9**(Suppl):1–15.
[5] Marks HG, Augustine P, Allen RJ. Fisher's syndrome in children. *Pediatrics* 1977;**60**:726–9.
[6] Kibel MA. Guillain-Barré syndrome in childhood. *S Afr Med J* 1983;**63**:715.

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Aetiology of growth hormone deficiency

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SUMMARY A retrospective analysis was performed in an attempt to identify perinatal risk factors for the development of growth hormone deficiency. More of the affected children were boys, and their birth weights were significantly lower than those of the national average; there were also considerably more preterm and post-term deliveries among boys.

The aetiology of idiopathic growth hormone deficiency[1] is multifactorial and there is an increased incidence of traumatic deliveries in affected children.[2 3] This study was undertaken to assess whether this association exists in the United Kingdom, and to attempt to identify other perinatal risk factors.

Methods

We studied cases of idiopathic growth hormone deficiency successfully submitted to the Health Services Human Growth Hormone Committee between January 1980 and June 1984 from 18 of the 19 regional growth centres in the United Kingdom. Details of affected patients were obtained from the growth hormone request forms then in use. Information abstracted included the age and sex of the patient, date of birth, gestation, mode of delivery, birth weight, heights and ages of parents. Statistical analyses were by the χ^2 and unpaired t tests. We used the population perinatal data that were collected for the National Perinatal Epidemiology Unit in association with the Office of Population Censuses and Surveys.[4] Anthropometric data were obtained from national sources.[5]

Results

We studied 300 children (196 boys and 104 girls). Of these, 275 children had isolated growth hormone deficiency and the remaining 25 also suffered from additional endocrine abnormalities. The mean (SD) age of boys was 10·4 (4·0) years and that of girls 9·3 (4·2).

No significant differences were found between affected children and the averages for the normal population concerning the heights of parents, ages of mothers or modes of delivery. The ratio of boys to girls in the sample was 1·88:1, which was significantly greater than the population ratio of 1·06:1 ($p<0·001$). The gestational age, available for 298 of the children, showed that boys with growth hormone deficiency had significantly more preterm (less than 36 completed weeks of gestation) and post-term (greater than 42 weeks of completed gestation) deliveries than the general population ($p<0·001$); this correlation was not found in girls.

The mode of delivery was started for 287 children. We were unable to examine breech deliveries separately, as the figures for England and Wales were not collated separately and the figures for Scotland, although available, were too small for analysis. There was, however, no significant increase in the proportion of instrumental deliveries or caesarean sections in the children deficient in growth hormone.

The birth weights of 290 of the children were recorded. The mean (SD) birth weight of boys was 3·18 kg (0·63) and that for girls 3·15 kg (0·62). If we consider the children delivered at term separately the figures rise to 3·29 kg (0·51) and 3·20 kg (0·47), respectively. Compared with national figures the children deficient in growth hormone who were delivered at term were significantly lighter ($p<0·001$). The 25 children with additional endocrine abnormalities were not significantly different from the remainder of the group.